



ISSN:  
Electronic version: 1984-5685  
RSBO. 2020 Jan-Jun;17(1):76-83

## Case Report Article

# Juvenile trabecular ossifying fibroma in association with multiple mandible deciduous teeth: an atypical case report

Rafael Correia Cavalcante<sup>1</sup>

Camila Lago<sup>2</sup>

Luiz Eduardo Baglioli<sup>2</sup>

Aline Costa Vaz<sup>2</sup>

Aimee Simonetto<sup>2</sup>

Mariana Dalledone<sup>2</sup>

Allan Fernando Giovanini<sup>2</sup>

Rafaela Scariot<sup>1</sup>

### Corresponding author:

Rafaela Scariot

Rua Prof. Pedro Viriato Parigot de Souza, n. 5.300 – Campo Comprido

CEP 81280-330 – Curitiba – Paraná – Brasil

E-mail: [rafaela\\_scariot@yahoo.com.br](mailto:rafaela_scariot@yahoo.com.br)

<sup>1</sup> Department of Dentistry, Federal University of Paraná – Curitiba – PR – Brazil.

<sup>2</sup> Department of Dentistry, Positivo University – Curitiba – PR – Brazil.

*Received for publication: October 6, 2019. Accepted for publication: November 11, 2019.*

### Palavras-chave:

fibroma; ossifying;  
fibrous dysplasia of  
bone; mandible.

## Abstract

**Introduction:** Ossifying fibroma is a benign fibro-osseous lesion characterized by the formation of a cellular fibrous connective tissue stroma with cementum-like material and/or osseous components. The juvenile variant is reported to be an aggressive lesion in paediatric patients. Its poor symptomatology contrasts with its highly aggressive behaviour in maxillary or mandibular region. **Objective:** The aim of the present study is to report a case of an atypical ossifying fibroma in association of multiple deciduous teeth. **Case report:** A nine years-old girl was referenced to the Oral and Maxillofacial Surgery Service at Positivo University by the Paediatric Service after radiographic confirmation of a round lesion in lower left mandible region. Imaginological examination revealed a well-defined, round, with multiple radiopaque foci associated with the roots of deciduous molars and with root of first permanent mandibular molar of the left side. Lesion embedded the root and crown of the unerupted second left premolar, suggesting that the tumour arose from it. After incisional biopsy, histopathological analysis suggested the trabecular pattern of juvenile ossifying fibroma. Surgery was conducted through

an intra-oral approach. Incision was drawn in gingiva from the left lateral inferior incisor region until the unerupted second molar region, finished with a relaxing incision. A round tumour in association with the unerupted permanent premolars, which were also extracted, was removed in its entire extent by enucleation. Histopathological analysis confirmed the previous diagnosis of juvenile trabecular ossifying fibroma. Peripheral ostectomy was conducted to diminish chances of lesion relapse. **Conclusion:** 16 months follow-up showed no lesion recurrence.

## Introduction

Ossifying fibroma is a benign fibro-osseous lesion characterized by the formation of a cellular fibrous connective tissue stroma with cementum-like material and/or osseous components [1-3]. This neoplasm was also known as cemento-ossifying fibroma and cementifying fibroma, however, in 2005 the new WHO classification reduced these terms to ossifying fibroma [4]. It has been suggested that ossifying fibroma arises from the periodontal membrane [9], and this can be stimulated by previous tooth extraction and periodontitis [5, 11, 14]. Ossifying fibroma is most prevalent in the mandible, especially in the premolar and molar regions [4, 12]. Patients in the second to fourth decades of life are the most affected. There is a predilection for women [3, 4]. MacDonald-Jankowski [10], in a systematic review, evaluated the principal features of 781 cases of ossifying fibroma. He found that females were more affected than males, and the ossifying fibroma was more prevalent in the mandible (3:1), in patients in fourth decade of life [10]. Juvenile ossifying fibroma (JOF) is a bro- osseous lesion caused by a rare type of tumour which appears most commonly before 15 years of age [1, 4, 10].

Clinically, most cases are asymptomatic with a slow and progressive growing [3, 17], swelling and facial asymmetry. Swelling is mainly the only complaint of this pathology [4, 19]. Usually, the ossifying fibroma is found in routine radiographic examinations [4, 19, 21]. In some aggressive cases, on the other hand, a destructive behaviour and facial asymmetry are observed [1, 4, 19, 21]. Juvenile ossifying fibroma is reported to be an aggressive lesion in paediatric patients. Its poor symptomatology contrasts with its highly aggressive behaviour in maxillary or mandibular region.

In imaginological exams, the ossifying fibroma appears as a round or ovoid expansive and well-

circumscribed lesion, with a thin radiolucent line [5, 12, 20]. The radiopacity is associated with the quantity of calcified material presented; early lesions appears as radiolucent areas, while mature as radiolucent mixed with radiopaque [12]. The teeth adjacent to the lesion may have their roots displaced, but root resorption is uncommon [2].

Histologically, ossifying fibroma presents a highly cellular fibrous stroma with cementum-like or bone masses, or both [6, 7, 19]. The cellular fibrous stroma is similar to the chinese-letter shape of trabecular in fibrous dysplasia [8, 9, 18]. Fibrous dysplasia and ossifying fibroma have histopathological similarities, and the clinical and radiographic findings are important for the differential diagnosis [15]. The association of clinical and radiographic findings with histopathological analysis is indispensable for the adequate management. JOF was subdivided in a report conducted by El-Mofty into two histopathological variants: the trabecular (TrJOF) and psammomatoid (PsJOF). One clinical feature that helps differentiate between the histopathological variants is the site of involvement; PsJOF is reported to be located mainly in the paranasal sinuses, and TrJOF mainly in the maxilla [6, 7, 15, 18, 19]. TrJOF features a proliferation of cellular fibroblastic tissue admixed with woven bone trabeculae with varying histologic presentations [18, 19]. A TrJOF was found to be unusual within the mandibular molar region.

The recommended treatment to ossifying fibroma is complete excision of the lesion by enucleation and curettage or resection [4, 14, 15]. En-bloc resection should only be considered in cases of relapse after curettage, aggressive behaviour or extensive erosion of multilocular lesion [2]. Ossifying fibromas treated by enucleation have been shown a low relapse [4]. Based on the aggressive JOF nature and its high rate of recurrence, some authors advocate performing a large mandibulectomy followed by reconstruction

of the mandibular defect by a micro-anastomosed osseous flap. These invasive procedures can lead to functional and aesthetic impairments in growing patients. Spontaneous bone tissue regeneration is an uncommon event, other than in the healing of fractures although there is the potential for bone regeneration in children after mandibular resection, particularly when the periosteal layer is spared.

Different cases of JOF have been reported in literature with different treatment modalities. The main purpose of the present paper is to report a case of an ossifying fibroma in a nine years-old patient associated with multiple deciduous teeth.

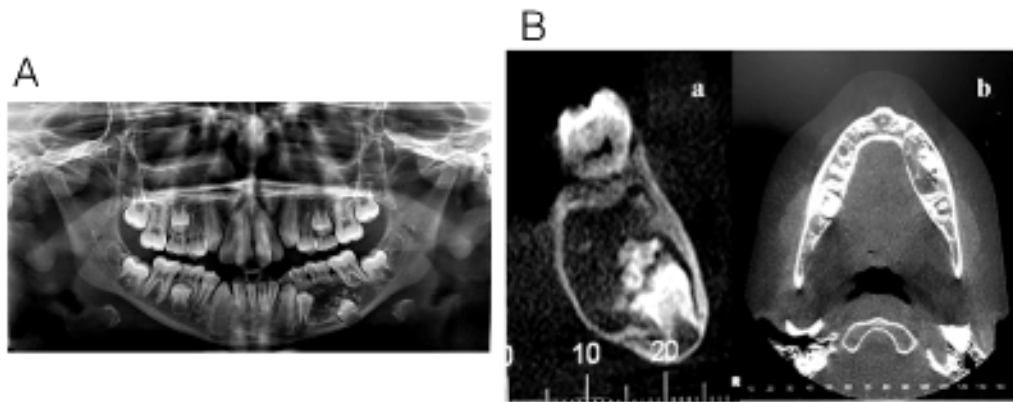
## Case report

A nine years-old girl was referenced to the Oral and Maxillofacial Surgery Service at Positivo University by the Paediatric Service after radiographic confirmation of a round lesion in lower left mandible region. Facial analysis presented no visible alterations (figure 1). Patient was systemically comprised with renal insufficiency (proteinuria and haematuria) association with Berger syndrome suspect. Intraoral examination showed no swelling, no pain, and no deciduous teeth mobility of those around the lesion.



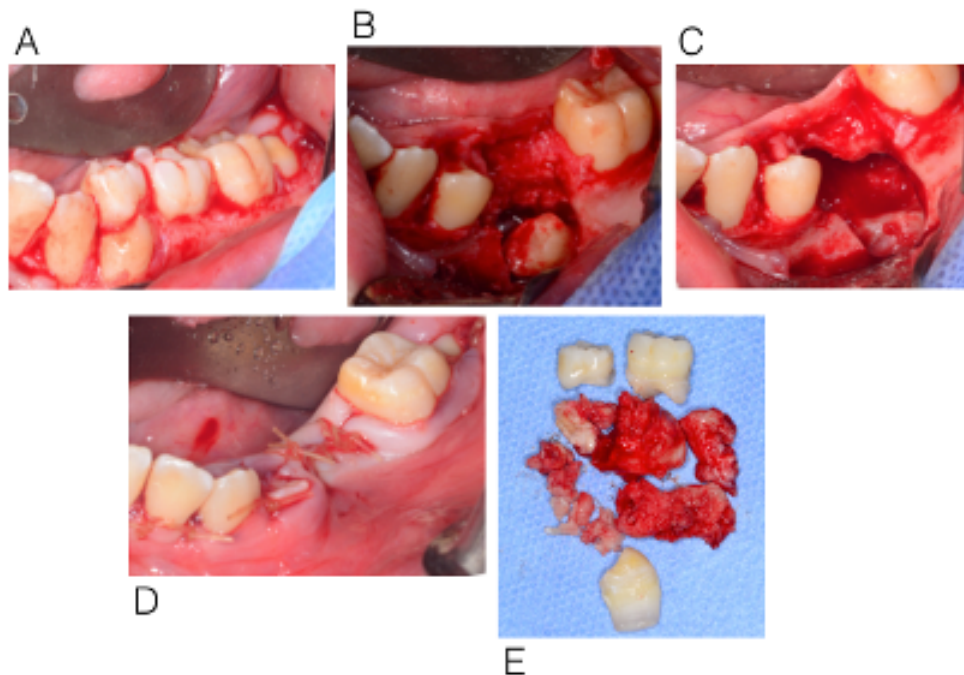
**Figure 1** – A) Frontal photograph of the nine-year old patient showing no visible facial asymmetry; B) and C) intrabuccal photographs. Intraoral examination showed no swelling, no pain, and no deciduous teeth mobility of those around the lesion

Panoramic radiography showed an intraosseous lesion associated with the crown and root of a no erupted premolar and with the roots of the deciduous molar teeth. Computed tomography revealed a well-defined, low-density mass associated with points of high-density in the left mandibular premolar-molar region (figure 2). Both the crown and the root of the unerupted first and second premolar were embedded in the lesion.



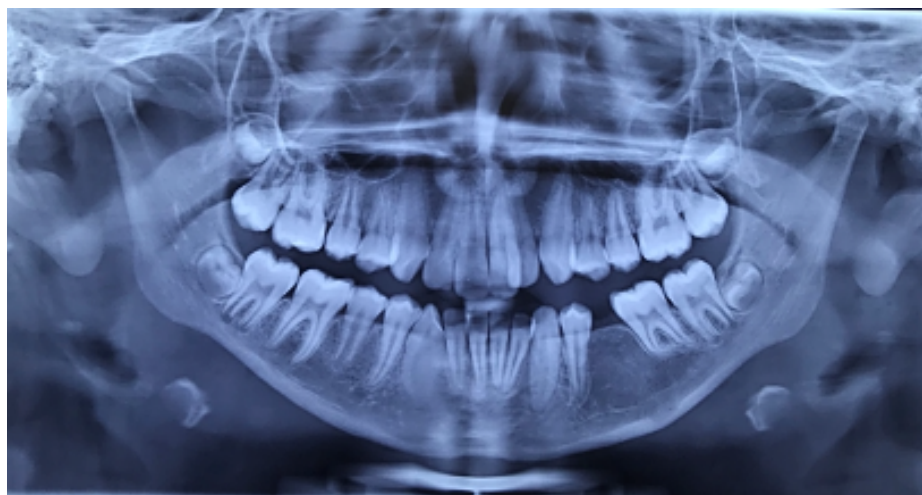
**Figure 2** – A) Panoramic radiograph showed an intraosseous lesion associated with the crown and root of a no erupted premolar and with the roots of the deciduous molar teeth; B) computed tomography showed a well-defined, low-density mass associated with points of high-density in the left mandibular premolar-molar region. Both crown and root of the unerupted first and second premolar were embedded in the lesion

An incisional biopsy was conducted with local anaesthesia to further evaluate the histological features of the lesion. The patient was then prepared to enucleation of the lesion under general anaesthesia. An incision was conducted in the gingiva from the left lateral inferior incisor region until the region of the unerupted second molar finished with a relaxing incision in the same region. A round tumour in association with the permanent premolars, which were also extracted, was removed. Peripheral ostectomy was conducted to diminish the chances of lesion relapse. Suture was then conducted after periosteum liberation. Lesion was referred to the oral pathology department to final biopsy and diagnosis confirmed ossifying fibroma (figure 3). Patient stayed in the hospital until the end of the day to finish intravenous analgesic therapy and it was dismissed with oral analgesics to continue controlling the pain. After 16 months follow-up, lesion presented no signs of relapse (figure 4).



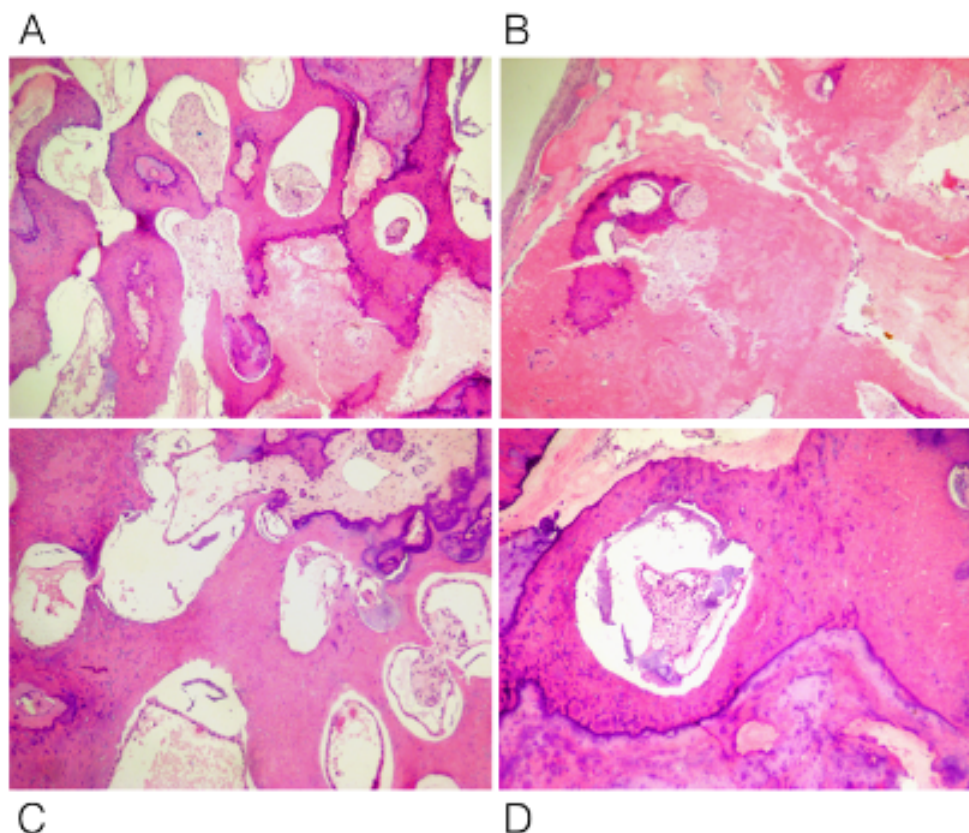
**Figure 3** – A) An incision was conducted in the gingiva from the left lateral inferior incisor region until the region of the unerupted second molar finished with a relaxing incision in the same region; B) a round tumor in association with the permanent premolars, which were also extracted, was removed; C) peripheral ostectomy was conducted to diminish the chances of lesion relapse; D) suture was then conducted after periosteum liberation; E) lesion was referred to the oral pathology department to final biopsy and diagnosis confirmed ossifying fibroma





**Figure 4** - Panoramic radiograph after one year and half follow-up showing no lesion relapse

Histopathological analysis showed spindle cell rich stroma admixed with immature inter-connecting osteoid trabeculae. Hypercellular immature osteoid trabeculae with numerous plump osteoblasts was also observed. The lesion presented itself well delimited but not encapsulated (figure 5).



**Figure 5** - Histopathological analysis showed A) and B) spindle cell rich stroma admixed with immature inter-connecting osteoid trabeculae. The lesion presented itself well delimited but not encapsulated. C) and D) Hypercellular immature osteoid trabeculae with numerous plump osteoblasts was also observed

## Discussion

Ossifying fibroma can be classified into two main types: the cemento-ossifying fibroma and the juvenile ossifying fibroma (JOF) [16]. The JOF is classified into two histological variants according to the World Health Organisation (WHO), the trabecular (TrJOF) and the psammomatoid (PsJOF) variants. JOF is usually seen in children and adolescents (mean age of presentation: 8.5-12 years old) whereas the later usually affects a wider patient age range (16-33 years-old). Both entities are relatively rare, however, PsJOF is more commonly encountered when compared to TrJOF [16, 17].

The enteropathogenesis of TrJOF is not well understood. GNAS or HRPT2 mutations have not been consistently found in TrJOF, which suggests that the etiopathogenetic of TrJOF differs from that of fibrous dysplasia [16-18]. Clinically, patients with TrJOF are often asymptomatic and early lesions are usually discovered as incidental radiographic findings. Displacement of teeth may be an early sign of this tumour process [19, 20]. TrJOF may be aggressive in its growth potential and as it matures, rapid growth may result in facial asymmetry and jaw deformity [2, 21].

Radiographically, an ossifying fibroma appears as a round or ovoid expansive and well-circumscribed lesion with a thin radiolucent line [5, 12, 20]. Ossifying fibroma attached to the tooth roots as well as in dental germ is an uncommon feature of this lesion, although this occurs in other lesion types. Computed tomography revealed a low-density mass associated with points of high-density [12, 13]. TrJOF tend to expand concentrically from a central point or epicentre, outward in all directions, and this expansion may result in displacement of teeth and the inferior alveolar nerve canal. Importantly the outer cortical plate remains intact despite significant expansion and thinning. Resorption of teeth is common. Unlike the ground glass, ill-defined blending borders in fibrous dysplasia, TrJOF maintains a well-defined corticated border [6, 20, 21]. The rapid progression of this neoplasm can mimic malignancy and osteosarcoma is suspected in younger patients, however; osteosarcomas radiographically display destructive irregular cortical margins invading the periodontal ligament and soft tissues, and do not have a thin radiolucent corticated boundary as seen in TrJOF [20].

The primary histologic criterion for JTOF consists of a neoplasm predominantly composed of cellular fibroblastic tissue with thin trabeculae of immature bone. This immature bone may

anastomose to form a lattice [16, 17]. TOF is usually well demarcated but unencapsulated. Of note, there may be considerable variation in stromal cellularity. Plump osteoblastic rimming of bone is a common feature. Clusters of osteoclastic multinucleated giant cells, areas of haemorrhage and foci of pseudocystic stromal degeneration may be observed [16]. Occasionally mitoses are seen in cell-rich areas. It has been suggested that rapid growth may be correlated with the secondary development of an aneurysmal bone cyst component [2, 16].

Differential diagnosis of TrJOF includes PsJOF, cemento-ossifying fibroma, fibrous dysplasia and osteoblastoma [6, 8, 22]. PsJOF differs from TrJOF in that it does not feature the thin trabeculae of immature bone as seen in TrJOF but is characterized by a proliferation of cellular fibroblastic tissue with a predominance of small basophilic ovoid concentric cementum-like spherules that feature peripheral brush borders that tend to blend into the connective tissue [6, 16]. Although cemento-ossifying fibroma is a histologic mimic of TrJOF, it notably occurs in older individuals. Fibrous dysplasia is characterized by irregular bony trabeculae with curvilinear shapes embedded in a fibrous background. It typically encompasses the osseous cortex. Cemento-osseous dysplasia (COD) displays a variable histologic picture of cellular fibroblastic tissue with deposits of woven bone, lamellar bone and cementum-like spherules depending on the stage of maturation. In contrast to TrJOF, the bony trabeculae in COD are thicker and less delicate and show less osteoblastic rimming. Furthermore, the cementum-like spherules in COD are unevenly shaped and display retraction from the stroma. Peripheral aggregates of haemorrhage are common in JTOF whereas in COD, haemorrhage is seen throughout. Osteoblastoma features a proliferation of plump osteoblasts within a vascular stroma and multiple layers of over-lapping osteoblasts and osteoclasts on more basophilic appearing bone [6, 16].

The recommended treatment to ossifying fibroma is complete excision of the lesion by enucleation and curettage or resection [4, 15]. The enucleations of ossifying fibromas have been shown a low relapse [4]. According to Liu *et al.* [9], if completely excised, most of ossifying fibromas do not recur. The prognosis for TrJOF is variable and recurrence following removal is not uncommon (~30-67% recurrence rate), especially if residual tumour persists following incomplete excision. There is no potential for malignant transformation of TrJOF. The lesion presented in this case report did not relapse after 1.5 years follow-up.

## Conclusion

Ossifying fibromas are uncommon in children. An association of the clinical and radiographic findings with the histopathological analysis is important for the correct diagnosis, and for the best management of the case.

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